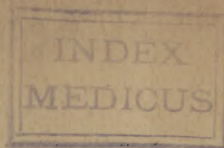


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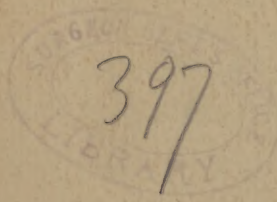
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MUSCULAR ATROPHIES.

BY

FREDERICK PETERSON, M. D.

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A CASE OF LOCOMOTOR ATAXIA ASSOCIATED
WITH NUCLEAR CRANIAL NERVE PALSIES
AND WITH MUSCULAR ATROPHIES.¹

By FREDERICK PETERSON, M.D.,

Attending Physician to the New York Hospital for Nervous Diseases.

THE case whose clinical history I am about to report has been a patient of mine at the New York Hospital for Nervous Diseases on Blackwell's Island since March 27, 1890. After having made a careful study of his condition, I discovered that he had been under the charge of Dr. E. C. Seguin at the Manhattan Eye and Ear Hospital from 1884 to 1886, and that Dr. Seguin had described the features of the case as they then existed in the JOURNAL OF NERVOUS AND MENTAL DISEASE for May, 1888. It is the first of five cases of ophthalmoplegia reported by the author in that journal.

There have been so many new developments in his condition during the past four years that I may be pardoned for briefly outlining his complete history from the first observations made to the time when he became a patient of my own.

M. J. T., now thirty-seven years of age, had a chancre and secondary symptoms fifteen years ago, that is, about 1875.

In 1882, he discovered, one morning, dimness of vision and external strabismus of the left eye with diplopia. A little later he had shooting-pains in the legs, occasionally in the arms.

In 1883, he had a momentary loss of consciousness, and fell, cutting his head. His left testicle also became swollen and hard during this year, and he was on specific treatment at Hot Springs for some time.

¹ Read before the American Neurological Association at Philadelphia, June , 1890.



In 1884, he had partial double ptosis. In the right eye, the internal rectus, inferior oblique, and sphincter iridis were paralyzed, and the superior and inferior recti feeble. The other muscles were normal. In the left eye the muscles supplied by the third nerve acted variably and feebly. The other muscles were normal. Both pupils were completely motionless to light and accommodation, the left larger than the right. There was overaction of the occipito-frontalis. Ophthalmological examination resulted as follows:

$$\text{Right V. } \frac{20}{70} + \frac{1}{18} = \frac{20}{40}.$$

$$\text{Left V. } \frac{20}{50} + \frac{1}{18} = \frac{20}{40}.$$

Accommodation right $\frac{1}{10}$, left $\frac{1}{14}$. No lesion of the optic nerves.

The left cheek was a little inactive, and there was a mild paresis of the right hand. (Dynamometer: R. 42—44, L. 45.) No Romberg symptom. Knee-jerks exaggerated. Both feeble and involuntary micturition.

In 1885, the ptosis was nearly total on the left, but partial on the right side. In the right eye the condition of the muscles was unchanged, while in the left they had improved so much that they acted almost normally. Some paresis and atrophy of both temporal and both masseter muscles were now noted.

In 1886, when lost sight of by Dr. Seguin, the ptosis was a little greater, the bladder still parietic, and the masticatory muscles unchanged. No marked facial paresis. The knee-jerks previously exaggerated had fallen to about normal.

Dr. Seguin, writing in 1888, said of this case:

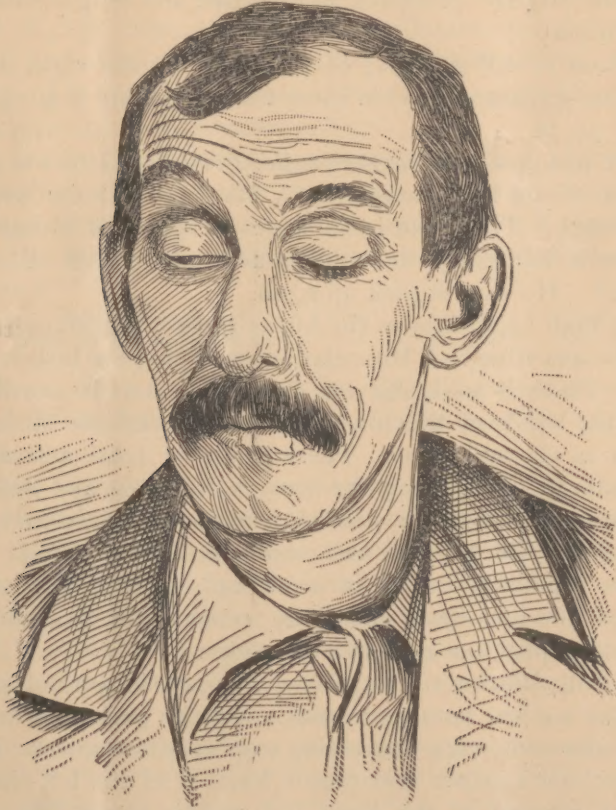
"The fulgurating pains, with hyperalgesia, and the fall in degree of knee-jerk, during two years of observation [1884-1886], would seem to justify a suspicion of incipient posterior spinal sclerosis."

Since 1886 until the present time there has been gradual progress in his disease. The condition of his eyes is now as follows:

In the right all of the muscles external and internal, except the rectus externus, are completely paralyzed. The rectus externus is parietic, and when moved exhibits clonic spasm. In the left eye there is almost complete ophthalmoplegia externa et interna, but the superior and inferior

oblique muscles still move a very little and very feebly. Both pupils are widely dilated, equal and immobile. Divergent squint of right eye, pulled outward by the parietic rectus. Accommodation paralyzed. Vision is unchanged.

As far, therefore, as the innervation of the ocular musculature is concerned, we have now lesions affecting both third nerves, both fourth nerves and both sixth nerves.



The weakness and atrophy of the masseter and temporal muscles are, if anything, more pronounced than before. In eating he has to support and assist the lower jaw with his hands. The two pterygoids on each side are also parietic. He cannot move the jaw forward or from side to side. The temporals and masseters do not react to faradism. The

wasting is evident in the photograph. There is no anæsthesia of the face. The left side of the face is a little paretic. It will be noticed in the photograph that the action of the occipito-frontalis is stronger and the nasolabial fold deeper on the right than on the left side. The tongue deviates slightly toward the left. The other cranial nerves are normal.

The electric reactions in the face and tongue muscles are normal.

There is still evidence of weakness in the right hand, for the dynamometer in three trials registers R 40-30-30, L. 35-35-38.

He has now well-marked *tabes dorsalis*. There is great ataxia of all four extremities. He cannot walk without assistance. The knee-jerks have disappeared altogether. There is numbness, anæsthesia and analgesia in all of his fingers. He cannot pick up a pin.

In both legs as far as the knees there is almost complete tactile anæsthesia. Muscular sense is entirely lost in both feet. There is analgesia and diminished tactile sensibility over the whole of the lower extremities and on the trunk as far as the umbilicus. There is almost total anæsthesia to heat and cold from the feet to the hips, and deferred sensation is a constant, and *allocheiria* an occasional phenomenon. The plantar and cremasteric reflexes fail.

He has incontinence of urine and the anal sphincter is weak. He has never had any crises. He has now no lightning-pain and no girdle sensation.

But in addition to the fully-developed locomotor ataxia and the nuclear paralyses mentioned, the patient now presents also some very interesting trophic disturbances. I will not include a rather remarkable fracture of the left clavicle at the junction of the inner and middle thirds (the outer portion is dislocated downwards one inch), for it is not apparently of recent date, and the patient never knew of its existence until I discovered it, and its history is therefore obscure.

Certain muscular atrophies are, however, very prominent. Besides that of the muscles supplied by the motor

branches of the two trigemini already described, there is conspicuous wasting of the right trapezius with a degenerative reaction.

In the left upper extremity there is complete paralysis of the two long extensors of the phalanges of the thumb, and atrophy and paresis of the abductor minimi digiti and of all of the interossei and lumbricales, with degenerative reaction. The left hand assumes almost the position of a *main en griffe* when at rest. The illustration of the two hands reveals the deformity only to a slight extent.

On the right side there is almost complete atrophy of the opponens and abductor pollicis.

Considerable wasting is apparent in the adductors of both thighs, more upon the right side; and there is a marked difference in the circumferences of both the thighs and the legs, as is evident from the following measurements:

Circumference of	Right	Left.
Thigh, 13 cm. above patella.....	40 -37	42.5-39
Leg 13 cm. below patella.....	29 5-28	27.5-26

From this it is clear that the right thigh is smaller than the left, whereas the left leg is smaller than the right by some two cm.

The electrical reactions are quite normal in all of the muscles of the lower extremities, except the adductors of the thigh and the sartorius of the right side, where the contractions to faradism are weak and "wabbling."

Most of the muscles of the body were examined with both the faradic and galvanic currents, and those in which partial or complete degenerative reaction was present are exhibited in the accompanying table.

ELECTRICAL REACTIONS IN ATROPHIED MUSCLES—CASE OF M. J. T.

Muscles.		Faradic (Primary).	Cm.	Galvanic.	No. of Grenet cells.
Left.	Ext. primi internodii pollicis.	No contraction.....	12	No contraction.	31
	" secundii "	" "	12	" "	31
	First interosseus.....	Slow "	10	Slight K C C....	31
	Second "	No "	12	No contraction.	31
	Third "	" "	12	An C C > K C C..	31
	Fourth "	Slow, wabbling contraction..	12	K C C > An C C..	18
	Abductor minimi digiti....	Slow contraction	12	K C C > An C C..	27
Right.	Trapezius.....	Contraction in parts....	12	A C C = K C C..	16
	Abductor pollicis.....	No contraction.....	12	A C C > K C C..	31
	Opponens pollicis.....	Slow, wabbling contraction..	12	A C C > K C C..	31
	Sartorius.....	" " "	12	A C C > K C C..	25
	Adductors of thigh.....	" " "	12	A C C = K C C..	25

Summary.—The main features of the case, then, may be summarized as follows: The patient has a number of bilateral motor-cranial nerve palsies, viz.: the third, fourth, fifth and sixth nerves; and for convenience of historical survey and clearness I have made use of Dr. Starr's diagram of the third nerve nuclei,² adding thereto very schematically the nuclei of the other affected nerves, and inserting the dates of their lesions, as pointed out in the clinical history.

SCHEMATIC REPRESENTATION OF NUCLEI OF NERVES AFFECTED
TO SHOW THE HISTORY MERELY.

Left.	Median line.		Right.
<i>Sphinct. iridis.</i> Paralyzed, 1884.	<i>Ciliary musc.</i> Paralyzed, 1890.	III.	<i>Ciliary musc.</i> Paralyzed, 1890.
<i>Levat. palp.</i> Feeble, 1884.	<i>Rect. int.</i> Feeble, 1882 to 1884.		<i>Rect. int.</i> Paralyzed, 1884.
Paralyzed, 1885.	Normal, 1885. Paralyzed, 1890.		<i>Levat. palp.</i> Feeble, 1884. Paralyzed, 1885.
<i>Rect. sup.</i> Feeble, 1884.	<i>Rect. inf.</i> Feeble, 1884.		<i>Rect. sup.</i> Feeble, 1884.
Normal, 1885. Paralyzed, 1890.	Normal, 1885. Paralyzed, 1890.		Paralyzed, 1890.
<i>Obliq. inf.</i> Feeble, 1884. Normal, 1885. Almost completely paralyzed, 1890.			<i>Obliq. inf.</i> Paralyzed, 1884.
<i>Obliq. sup.</i> Almost completely paralyzed, 1890.		IV.	<i>Obliq. sup.</i> Paralyzed, 1890.
<i>Temp.</i> Feeble, 1885. Feeble, 1890.	<i>Masseter.</i> Feeble, 1885. Feeble, 1890.	V.	<i>Masseter.</i> Feeble, 1885. Feeble, 1890.
<i>Pteryg. ext.</i> Feeble, 1890.	<i>Pteryg. int.</i> Feeble, 1890.		<i>Pteryg. int.</i> Feeble, 1890.
	<i>Rect. ext.</i> Normal, 1884. Normal, 1885. Paralyzed, 1890.	VI.	<i>Rect. ext.</i> Normal, 1884. Normal, 1885. Feeble, 1890.

The patient has also exhibited slight traces of a crossed paralysis for four or more years. He has, furthermore, locomotor ataxia, as shown by the occurrence at one time

² Ophthalmoplegia Externa Partialis. By M. Allen Starr, M.D., Journal of Nervous and Mental Disease, May, 1888.

of lightning-pains and by the presence now of ataxia, widely-distributed anæsthesias, failure of knee-jerks, and ocular, vesical and anal symptoms.

Finally, he presents marked trophic changes in numerous muscles.

Pathology.—As to the morbid processes which underlie these various manifestations, there is, in the first place, undoubtedly a sclerosis of the posterior columns of the spinal cord.

The ophthalmoplegia is, of course, nuclear. Read in one way the symptoms on the side of the cranial nerves, taken in conjunction with the muscular atrophies and paralyses elsewhere, certainly very closely resemble the syndrome so well described by Dr. Sachs in his paper before this Association last year, under the title of *Polioencephalitis Superior and Poliomyelitis*.³ There is no reason to suppose that polioencephalitis superior and chronic poliomyelitis could not occur in combination with a posterior sclerosis, and it is by no means certain that the case described by Dr. Sachs may not ultimately develop locomotor ataxia, although the absent knee-jerks in both legs and the vesical and anal symptoms in his case may be explained on other grounds. On the other hand, however, these nuclear palsies, more especially of the motor nerves of the eye, are so common in *tabes dorsalis*, and the investigations of Déjerine,⁴ Nonne⁵ and others have demonstrated that muscular atrophy is not infrequently associated with locomotor ataxia. Déjerine reports 11 cases in 106, and in five of these he made a histological examination, finding the anterior horns normal, but a degenerative neuritis in the affected peripheral nerves. The most important matter to be settled in this case is whether the muscular atrophies are due to peripheral or central lesions. Speculation upon the question would seem to be of very little utility, and its solution must be left to the hoped-for autopsy.

³ Amer. Jour. Med. Sciences, Sept., 1889.

⁴ Gazette méd. de Paris, March 10, 1888, and paper before Société de Biologie, 1889.

⁵ Arch. f. Psych., vol. xix.

It has been assumed by a number of authors that total paralysis of all the muscles supplied by the third nerve implies not a nuclear, but a nerve-trunk palsy. Thus Dr. Starr⁶ says: "If all the muscles of the eyeball supplied by the third nerve are affected, *including the iris*, the case is one of total peripheral paralysis of the third nerve, and the lesion lies on the base of the brain, and may in time implicate other cranial nerves."

In my case all the muscles of both third nerves are totally paralyzed, including both irides, and yet there is every reason to believe that the palsies are nuclear. It would at least be difficult to conceive of a lesion at the base of the brain so widely and so symmetrically distributed as to affect the trunks of both third nerves, both fourth nerves, both sixth nerves, and the motor portions of both trigemini, yet permitting the escape of the sensory portions of the latter.

My own diagnosis of the disease from which the patient suffers is indicated by the title of this paper.

